

DISLOCATION OF THE HIP IN MYELOYDYSPLASIA

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THE orthopedic surgeon has a distinct responsibility together with the pediatrician, the neurosurgeon, and the urologist, in the care of children with myelodysplasia. Although little information has been published in regard to orthopedic manifestations of this entity, considerable improvement has been made in recent years in the management and rehabilitation of these children.

The extent of monoplegia or paraplegia varies from one patient to the next. There may be partial sparing of nerve fibers, allowing some innervation distal to the lesion, or there may be total paralysis beyond the level of the lesion. Close observation of these patients over some years may disclose that increased innervation has occurred. This improvement I interpret to be the result of increasing maturity of the spinal cord.

In the patient with flail lower extremities it is not uncommon to find bilateral dislocation of the hips as an acquired phenomenon. The mechanism of the dislocation is not fully understood, although two factors undoubtedly are contributory. One is the tendency for the child with this disorder to keep the feet together, thus adducting both hips; because of paralysis, this constant positioning is unopposed by any abduction, which is the position of greatest stability for the hip. The other factor in production of hip dislocation frequently present in this situation is an active iliopsoas muscle, which is innervated from the second, third, and fourth lumbar nerve roots. Since the iliopsoas muscle inserts into the lesser trochanter, it exerts an anterior pull on the head and neck of the femur. Thus, the resting position of adduction plus the unopposed voluntary activity of the iliopsoas muscle results in a force that tends to displace the femoral head from the acetabulum.

Occasionally a unilateral dislocation occurs in these patients. If the resultant paralysis leaves only one lower extremity flail, this dislocation will occur as just described. There is another situation in which the hip abductors (gluteus medius, gluteus minimus, and tensor fasciae latae) are active on one side, while the opposite hip has little or no abductor activity. In this situation, the limb with good abductor power will assume the abducted position, while the opposite limb will seek a parallel position, thus creating an adducted hip. This situation sets the stage for dislocation.

Since these dislocated hips most often are acquired when the infant is six months of age or older, they do not represent the same problem as a congenitally dislocated hip. The acetabulum has sufficient depth for the femoral head, provided that the hip has not been dislocated a long time. When the dislocation has been present for some months, the acetabulum becomes shallow, a condition that will foster a somewhat insecure hip when it has been relocated.

Diagnosis

The method of diagnosis of a dislocated hip or hips in the myelodysplastic child is the same as that for congenital dislocation. Abnormal skin folds from the adductor muscle mass, clicking with flexion and abduction of the hip, absence of the femoral head in the femoral triangle, a palpable femoral head posteriorly, and instability with the "push-pull" technic in the involved limb—all support a provisional diagnosis of dislocation of the hip. Because one often finds a tight adductor muscle mass in the myelodysplastic child, the abduction and flexion test alone may not be diagnostic.

The roentgenogram will aid in confirming the diagnosis; there will be evidence that the dislocated femoral head is resting laterally and frequently superiorly to its usual position in the acetabulum (*Fig. 1*). Evidence of a break in the normal smooth

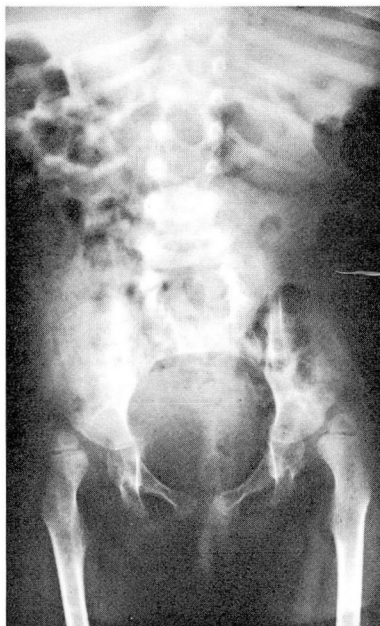


Fig. 1. Roentgenogram of 3-year-old girl with myelodysplasia and bilateral dislocation of the hips. Note spina bifida of lower lumbar and upper sacral vertebrae. A meningocele had been repaired one year previously.

arc outlined by the inferior cortices of the pubis and the femoral neck (Shenton's line) is also seen. There may be increased obliquity of the acetabular roof (the normal angle between the superior acetabulum and a line drawn between the two triradiate cartilages is 27 degrees) if the hip has been dislocated for several months. However, this is not obligatory and depends upon the age of the child at the time dislocation occurred, as well as upon the length of time that the femoral head has

been dislocated. A true lateral roentgenogram of the pelvis can also be taken to show the positions of the femoral heads in relation to the iliac brim of the acetabulum¹ (*Fig. 2*).

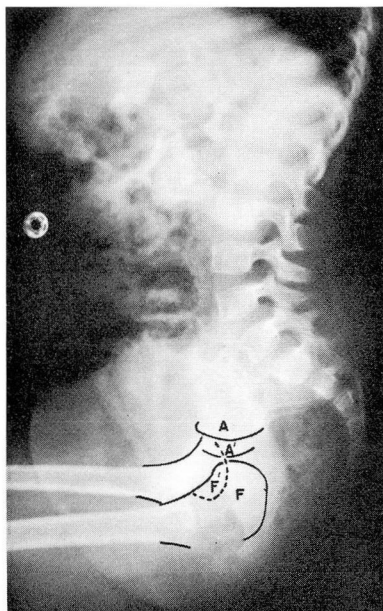


Fig. 2. Lateral roentgenogram of the pelvis. The femoral head (F) should rest in contiguity with the acetabulum (A). F¹ indicates the contralateral femoral head, and A¹ the corresponding acetabulum.

Treatment

The initial treatment that I prefer to use is reduction of the hip or hips on a Koontz traction and abduction frame (*Fig. 3*) followed by application of a bilateral Spica cast. This cast is changed every six weeks over a total cast-immobilization period of three months. At the end of the three months' immobilization with the cast, the hip or hips are examined for stability. If the hips seem stable I then change to bracing to maintain abduction. This can be done with an abduction brace similar to the Ilfeld brace,² or with the Ponseti³ bar, which is an adaptation of the Dennis-Browne night kicker. The Ponseti bar has a long semicircular bar attached to the soles of shoes. With casting and also with bracing one must constantly be on the alert to prevent localized pressure ulcers in these anesthetic limbs.

The stability of these hips should be reevaluated frequently after reduction. At that time there may be some motor power that was not present earlier in the involved extremities. If the hips appear stable, or if the improved motor power

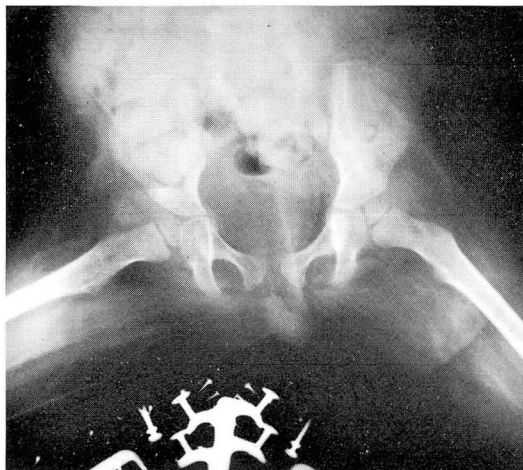


Fig. 3. Roentgenogram of the pelvis following reduction of the hips through use of the Koontz splint.

increases stability, then proper bracing and physical therapy can be instituted with the ultimate goal of walking with or without crutches. However, if stability is absent, or if subsequent instability is demonstrated, then one must plan for operative procedures to aid in securing the stability necessary for adequate ambulation.

One of the best operations to achieve stability in these hips is the innominate osteotomy described by Salter.⁴ This procedure brings the roof of the acetabulum into a more horizontal position, and swings the anterior rim of the acetabulum laterally. This operation effectively increases the depth of the acetabulum in the anterior and superior aspects; and thus improves the stability of the acetabulum in maintaining its relation with the femoral head. This operative procedure alone may add sufficient stability to the hip to allow ambulation.

However, some of the hips will still tend to become dislocated. In this instance, a Mustard⁵ transfer of the iliopsoas tendon from the lesser trochanter to the greater trochanter through a window in the ilium will create an abductor out of the iliopsoas muscle. Muscle testing to determine the strength of the iliopsoas muscle should precede such an operation. Hayes⁶ has been using this procedure for several years, and has observed much improved stability of the hip following such a transfer; the long-term results of this operation are still under observation. This accomplishes the removal of the iliopsoas muscle as a dislocating force, and converts it into a stabilizing abduction force. Hayes emphasizes the requirement of innervation at least as low as the third lumbar segment in order to achieve adequate power to maintain reduction.

As soon as a stable hip has been obtained, an active rehabilitation program should be instituted. Bracing will be necessary in most of these patients, and will

vary from long-leg, double upright braces attached to a pelvic band in a child with a severe involvement, to a short-leg, single upright brace for the patient with little residual paralysis. If a child who has dislocation of the hip in myelodysplasia can eventually be aided to achieve independence, the length of time and the effort involved in this process are well worthwhile.

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